



Diagnosing headache

BACKGROUND A systematic approach to the diagnosis of primary and secondary headache disorders requires the measurement of the frequency and the duration of headache.

OBJECTIVE This article presents thumbnail sketches of the most important headache subtypes and discusses some of the new revisions and headache types included in the International Classification of Headache Disorders (ICHD-2).

DISCUSSION The clinical features that distinguish secondary headaches requiring urgent investigation (red flags) from those that can be more safely monitored (blue flags) are discussed.

The clinician should approach the diagnosis of headache in an orderly, logical fashion. This will avoid the frustration of incorrect diagnosis and consequently poor treatment results. It is fortunate that headache is, in many of its facets, measurable. Before the 1988 International Classification of Headache Disorders (ICHD-1),¹ the criteria for making headache diagnoses were based on rather vague generalised symptom descriptions for each headache type. The 1988 classification centres on the duration and frequency of headache. The revised criteria of the 2004 edition (ICHD-2) form the basis for clinical diagnosis, treatment, and research.²

Primary headache

The diagnosis of a primary headache requires that all other identifiable and probable conditions that can secondarily cause headache be excluded. Therefore to make the diagnosis of primary headache, neither the history nor the physical examination should suggest a secondary cause for headache, and where appropriate,

special investigations such as computerised tomography (CT) of the head should have excluded such a condition.

For this crucial diagnostic step to take place, the clinician must be familiar with the typical features of each headache type, carried out a competent physical examination, and selected the appropriate special investigations. A simplified, general operational classification of headache that allows for the differentiation of primary and secondary headaches (but at an early stage divides primary headaches on the basis of frequency) is shown in *Table 1*.

Secondary headache

According to lifetime prevalence studies of headache, both primary and secondary tension type headache is the most common (69%), while headache from systemic infection is second in frequency (63%). Migraine is next (16%), followed by headache after head injury (4%), idiopathic stabbing headache (2%), exertional headache (1%), vascular disorders (1%), sub-arachnoid haemorrhage (<1%), and brain tumours (0.1%).³

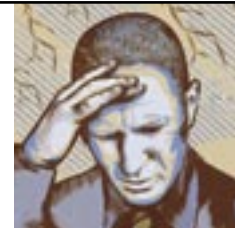
Secondary headaches are an important consideration, not only because of the potential danger inherent in some, but because of their prevalence. A clinical classification of secondary headaches is shown in *Table 2*.

Blue and red flag headaches

Blue and red flag headaches are those in which some feature of the history or examination suggests that the headache is due to a secondary cause. Blue flag features (*Table 3*) indicate secondary headaches that do not require urgent investigation. Red flag features require urgent attention (*Table 4*).

Diagnosis of primary headache syndromes

Once the clinician is confident that a secondary cause



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for headache has been excluded, the next orderly steps in the diagnostic algorithm can take place.

Frequency

Divide primary headaches into: low to moderate frequency (<15 headache days per month) – ‘episodic’ headaches, and high frequency (>15 headache days per month) – ‘chronic daily’ headaches.

Attack duration

Headaches can be readily categorised into two groups: those that last for more than 4 hours (eg. migraine), and

those that last less than 4 hours (eg. cluster). Using the primary distinguishing point of headache frequency per unit time, all headache patients can be categorised into two broad groups: episodic or frequent. These groups can be subcategorised into headaches of shorter or longer duration.

Episodic headaches

- Low to moderate frequency headache with longer duration of attacks (>4 hours per day): migraine, episodic tension type headache
- Low to moderate frequency headache with short duration of attacks (<4 hours per day): cluster headache and other trigeminal autonomic cephalalgias (TACs)
- Shorter duration primary headaches with special features (<15 days per month): headaches triggered by straining, coughing, Valsalva manoeuvre or sleep (primary cough, primary exertional, hypnic, orgasmic and pre-orgasmic headaches). Here the trigger is identified and the duration of the headache varies from being short acting to long lasting, possibly for days. Short stabbing headaches can also occur less than 15 days per month.

Clinical features

Head pain

Typical head pain of migraine is a unilateral, moderate to severe throbbing headache – often frontal in location – but rather diffusely compared to cluster headache. It is aggravated by positional change, exercise and jolting. It may change sides (side shift) from attack to attack or within the same attack.

Tension type headache is not so severe (mild or moderate), usually bilateral, described as ‘tight, gripping, pressing or aching’. It is not generally aggravated by positional change, exercise or jolting.

The head pain in cluster headache is excruciating, unilateral, located behind one eye, boring in character and not affected adversely by exercise or position. It may rarely shift from one attack to another to the other side, but tends to be ‘side locked’ for an individual.

Associated features

Seldom are there associated features in tension type headache, but in migraine and cluster headache, the associated features are frequent and useful in clinching the diagnosis (Table 5). Migraine associated features differ markedly from those of cluster headache.

Most migraine patients are nauseated and at least a third will actually vomit during an attack. This is not

Table 1. Primary and secondary headache disorders

Primary episodic headache disorders

- Migraine
- Episodic tension type headache
- Cluster headache and trigeminal autonomic cephalalgias

Primary frequent (chronic daily) headache disorders

- Transformed migraine
- Chronic tension type headache
- New daily persistent headache
- Hemicrania continua

Secondary headache disorders

Table 2. Secondary headaches

- Headache secondary to disorders of the:
 - skull (eg. Paget disease, mastoiditis, secondary malignancy)
 - ears (eg. otitis media or externa)
 - eyes (eg. glaucoma, strabismus, ocular strain, iritis)
 - nose and nasal sinuses (eg. acute or chronic sinusitis)
 - teeth (eg. tooth abscess, malocclusion)
 - cervical spine (eg. cervical spondylosis)
 - cranial nerves (eg. herpes zoster, occipital neuralgia)
- Headache secondary to:
 - intracranial vascular disorders (eg. venous sinus thrombosis, ruptured aneurysm, cerebral haemorrhage)
 - extracranial vascular disorders (eg. carotid artery dissection, cranial arteritis or carotidynia)
 - disorders of intracranial pressure (raised intracranial pressure or low cerebrospinal fluid pressure, eg. postlumbar puncture leak)
 - intracranial infections (eg. encephalitis or meningitis)
- Headache attributable to:
 - generalised infection (eg influenza, typhoid or malaria)
 - medication (eg. vasodilators)
 - hypertension

so in cluster headache. Patients in a migraine attack are frequently photophobic and dislike noise. They will actively seek darkness and avoid exposure to noise. Migraine sufferers may, either preceding or within the headache phase, experience aura symptoms. These are predominantly visual, although there may be numbness and paraesthesiae circumorally (often the upper limb), and sometimes difficulty in finding words, or clumsiness and weakness of a limb (usually the upper limb). These neurological changes will have a gradual onset and last for about half to one hour.

In cluster headache, there is usually conjunctival injection, lacrimation, nasal blockage and nasal discharge – all on the side ipsilateral to the headache. There is little nausea and very seldom vomiting.

Patient behaviour and appearance during attack

In typical migraine attacks, the patient lies down, chooses silence, tries to sleep (often finding that sleep relieves the headache), and does not want to be disturbed. Unfortunately, this is similar to patients with meningitis, encephalitis or subarachnoid haemorrhage.

The cluster headache sufferer – by contrast – cannot lie quietly, gets up and walks in an agitated fashion, and sometimes will beat their head or take up unusual positions of the body. Some sufferers run to ease the pain.

Onset and offset of the attacks

Migraine patients rarely have abrupt onset of their attacks. Many migraine sufferers will have prior warning of an attack in the form of a prodrome that occurs 24–48 hours before the aura or headache commence. The actual onset of events in migraine is slower; generally hours.

In cluster headache, the onset is typically abrupt, with an upswing of about 5 minutes to the peak of discomfort, a plateau of about half an hour, and then a gradual decrement of about 20 minutes. Tension type headaches are similar to migraines in their slower onset.

Duration of attacks

Untreated or unsuccessfully treated migraines last 4–72 hours. Cluster headaches are shorter, having a duration range of 15–180 minutes. Episodic tension type headache has a duration of 30 minutes to 7 days.

High frequency headache

Clinical features

The main categories of high frequency headaches (>15

headache days per month) with long duration are:

- chronic migraine (previously termed ‘transformed migraine’)
- chronic tension type headache
- new daily persistent headache
- hemicrania continua.

Table 3. Blue flag headaches

- Headache that is mainly occipital, but sometimes radiates to the temple, exacerbated by examination of neck mobility (cervicogenic headache or cervical spondylosis)
- Headache temporally linked to whiplash injury of the neck
- Headache related to reading (eye strain)
- Headache clearly temporally linked to the ingestion of medications (eg. vasodilators)
- Headaches associated with systemic viral illness (eg. influenza)

Table 4. Red flag headaches

- New onset, specific setting. New headache in the setting of cancer (metastases), HIV infection (opportunistic infection), postmanipulation or trauma of the neck, or associated with mild head trauma in the elderly (subdural haematoma)
- New headache that is persistent¹
- Focal signs or symptoms (other than the typical visual or sensory aura of migraine)
- Headache with focal neurological signs that precede or outlast the headache (the rare exception is hemiplegic migraine³)
- Headache that is progressive (may suggest a mass lesion)
- Headache of sudden onset (may indicate a bleed either into the subarachnoid space or the cerebral parenchyma)
- Headache with rash (may indicate meningococcal meningitis or Lyme disease)
- Persistent unilateral temple headache in adult life (may indicate cranial arteritis)
- Headache with a raised erythrocyte sedimentation rate (ESR) (may be an indication of cranial arteritis, collagen disease or systemic infection)
- Headache with papilloedema (raises the suspicion of raised intracranial pressure due to a mass lesion or benign intracranial hypertension)
- Nonmigraine headaches in pregnancy or postpartum (cerebral vein thrombosis can occur during or just after pregnancy)
- Headache triggered by cough or straining (may be an indication of either a mass lesion or a subarachnoid bleed¹)
- Headache clearly triggered by changes in posture (may indicate low cerebrospinal fluid [CSF] pressure, for instance due to spontaneous CSF leak)
- Headache associated with pressing visual disturbances (may indicate conditions such as glaucoma or optic neuritis)
- Headaches that have primary characteristics, but with unusual features

Shorter duration primary headaches with special features (as described on *Page 622*) may also occur on more than 15 days per month.

High frequency, short duration headaches not triggered by exertion, sexual activity or sleep are:

- episodic and chronic cluster headache
- episodic and chronic paroxysmal hemicrania
- short lasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndrome.

Short stabbing headaches can also occur on more than 15 days per month.

Chronic migraine

The term ‘chronic daily headache’ is a syndrome that indicates headaches present on more than 15 days per month, for more than 4 hours a day, for at least 3 months.⁶ The ICHD-1 proved inadequate to classify chronic daily headache sufferers, hence the Silberstein and Lipton (S-L) classification was devised and is widely used.⁴ What was termed ‘transformed

migraine’ in the S-L classification has been accepted in the ICHD-2 as ‘chronic migraine’. There is some controversy regarding the exact definition of chronic migraine. Clinical features include headache on more than 15 days per month, and a history of migraine with a gradual build up of headache frequency with the intermittent migraines finally coalescing into a chronic daily headache pattern punctuated by peaks of acute headache activity that have features of migraine (eg. throbbing unilateral headache, nausea, vomiting, light and sound sensitivity).³ Rarely, the migrainous features disappear, and only a chronic daily nonspecific headache is left.

Chronic tension type headache

Patients with chronic daily headache are seen less frequently than chronic migraine patients. Features are similar in that there is a history of gradually escalating headache frequency, coalescing into a chronic daily headache pattern, but there is no history or features of migraine. These intermittent, and later, chronic daily headaches have the features of tension type headache. The typical patient will have daily dull bilateral headaches characterised by a pressing or gripping or band-like character with no migrainous features.

New daily persistent headache

In this headache type, the crucial point is that the headache does not evolve from an intermittent episodic headache pattern, but begins at one time and simply continues as a daily headache, ie. there is no progression from either intermittent episodic tension type headaches or intermittent migraines into a chronic daily headache pattern. Usually, there are few migraine features present with features of chronic tension type headache. This is compatible with the ICHD-2 definition of new daily persistent headache (NDPH), however the S-L classification allows diagnosis as NDPH if migraine features are prominent as long as the condition commenced abruptly.⁴ There are few series documenting the exact clinical features of this condition,⁵ but in the author’s experience, the headache is moderate in severity, fluctuates in intensity from day-to-day and even within the same day, sometimes changes location from day-to-day, is sometimes punctuated by sharp jabbing pains, and is associated frequently with lethargy. It is more frequent than chronic tension type headache in tertiary referral centres.

Table 5. Clinical features of episodic headache

Characteristics of migraine

- Female predilection
- Often commences in adolescence
- Often a positive family history
- Associated visual, sensory, motor or cortical symptoms
- Unilateral throbbing headache
- Associated gastrointestinal symptoms of nausea and vomiting
- Associated light and sound sensitivity
- Duration of days
- Often recurrent, predictable triggers

Characteristics of tension headache

- Female predilection not so pronounced as in migraine
- Often begins in young adulthood or later
- No aura
- Tight band-like pressing headache
- No features of gastrointestinal disturbance, phono- or photo-phobia
- Unusually not as incapacitating as migraine or cluster headache

Characteristics of cluster headache

- Male predilection
- Severe, often excruciating pain
- Unilateral with seldom side shift
- Location of pain typically behind the affected eye
- Shorter duration
- Ipsilateral autonomic phenomena
- Seldom triggers (except for alcohol during a cluster)

Hemicrania continua

Hemicrania continua is a rare, strictly unilateral headache sometimes evolving from an intermittent pattern, and sometimes arising de novo, that has the features of cluster headache. The severity of the pain is usually moderate, but there can be severe pain during exacerbations. The autonomic features are not as pronounced as in cluster headache. It is dramatically responsive to indomethacin. Rarely, bilateral features and side shift have been documented.

The ICHD-2

The following headache forms are more clearly defined in the ICHD-2 classification.

Trigeminal autonomic cephalalgias

In recent years, it has become clear that cluster headache is one of a family of primary headache disorders characterised by both trigeminal and parasympathetic activation.⁷ These include cluster headache, paroxysmal hemicrania, and SUNCT syndrome. All feature recurrent attacks of orbitotemporal pain associated with parasympathetic features such as those found in cluster headache. The SUNCT syndrome is unique in that there are very frequent attacks (3–200) per day, with a dramatic onset and associated profuse tearing. Chronic paroxysmal hemicrania (CPH), has features of cluster headache, but the attacks are frequent (more than 5 per day) and shorter lasting (2–30 minutes). It is responsive to indomethacin. The ICHD-2 includes an episodic form of CPH.

Primary stabbing headache

This headache is characterised by recurrent stabs of sharp, jabbing pain in the distribution of the first division of the trigeminal nerve. These recurrent attacks are alarming for the patient and very often stops them 'in their tracks'. There are no autonomic features.

Hypnic headache

Hypnic headache occurs primarily in the elderly, is relatively short lived (~30 minutes) and wakes patients from sleep, often in the early morning hours. It is usually bilateral, and lacks the severity of cluster headache. There are no autonomic features.

Headache related to cough, exertion, or sexual activity

These headaches are important in that, although easily recognisable, structural pathology such as Arnold-Chiari

malformations, and vascular lesions such as aneurysms or arterial dissection should be excluded.

Summary of important points

- Exclude secondary headaches (in particular, spotting the red and blue flags).
- Divide the primary headaches into episodic and chronic daily headache on the basis of their occurring on less or more than 15 days per month.
- Divide them further into headaches that last longer or shorter than 4 hours.
- Consider the triggers.

Resource

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